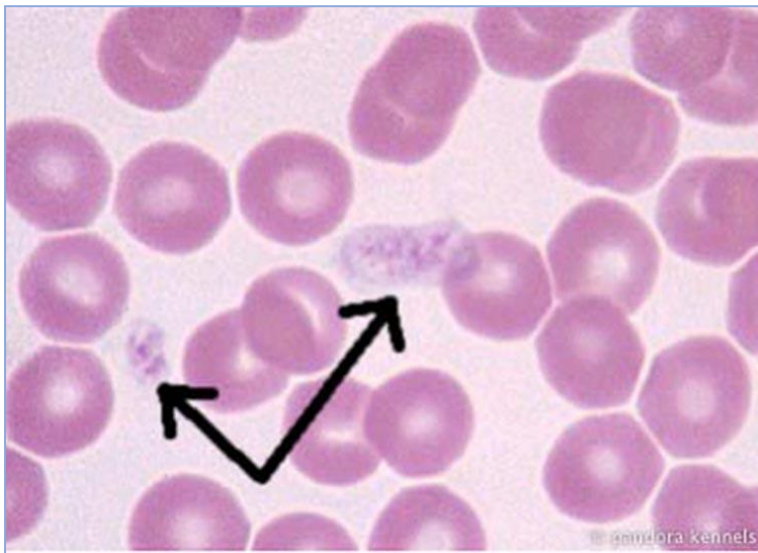


Von Willebrand's Disease

Most people are familiar with hemophilia A, an inherited blood clotting defect in human beings affecting only male children. Von Willebrand's disease is also an inherited blood clotting defect and dog breeds at high risk should be screened before being allowed to breed.

Breeds routinely tested are Doberman Pinscher, Golden Retriever, Shetland Sheepdog, Rottweiler, Miniature Schnauzer, German Shepherd, German Short-Haired Pointer, Standard Poodle, and Scottish Terrier.



Arrows point to two platelets amid a group of red blood cells

What is Von Willebrand's Factor?

Von Willebrand's factor is a protein complex produced both by platelets = blood cells involved in clotting and by the cells lining blood vessels. Von Willebrand's disease results when there is a defect in any one of these proteins. When a blood vessel tears and bleeding occurs, platelets are called to the area to clump upon each other, thus plugging up the hole and staunching the bleeding. Von Willebrand's factor acts as glue holding the platelets together and holds them onto the surface of the torn blood vessel.

When there is something wrong with one's von Willebrand's factor, platelets do not stick together properly and inappropriate prolonged wound bleeding occurs.

Types of Von Willebrand's Disease

There are three types of von Willebrand's disease.

TYPE I

In Type I, all the proteins making up von Willebrand's factor are present but only in very small amounts. This is the type common in the Doberman Pinscher, the Shetland Sheepdog, the German Shepherd Dog, and the Standard Poodle.

TYPE II

In Type II, the larger proteins making up von Willebrand's factor are completely absent, leaving only the smaller proteins to do the job. This creates more severe bleeding episodes and represents the type of von Willebrand's disease usually seen in German Short-Haired and Wire-Haired Pointers.

TYPE III

In Type III, there is simply no von Willebrand's factor at all. This is the most severe form and is usually seen in Scottish Terriers, Chesapeake Bay Retrievers, and Shetland Sheepdogs.

Von Willebrand's disease is not limited to the breeds listed here; forms of von Willebrand's disease have been found in over 50 breeds.

Unlike the genetics of hemophilia A in humans, as a sex-linked recessive trait, von Willebrand's disease is not as simple. Males and females are equally affected and the inheritance seems to be recessive but complicated.

Diagnosis of Von Willebrand's Disease

DNA testing is currently available. A swab of the inside of the cheek is all that is required to determine Von Willebrand's disease. There are 3 outcomes possible:

Clear = free of the disease

Carrier = carries the disease but is not affected by the disease

Affected = dog will have Von Willebrand's disease

Treatment of the Affected Dog

When hemorrhage is occurring or is anticipated (as with a planned surgical procedure), the best treatment is administration of von Willebrand's factor by transfusion. Pure von Willebrand's factor cannot be purchased from a blood bank but a blood product called cryoprecipitate, which is particularly rich in von Willebrand's factor, can be. Complete plasma is the next best choice and is much more available than cryoprecipitate. Administration of cryoprecipitate improves bleeding time for approximately 4 hours after administration.

A hormone called DDAVP (or desmopressin acetate) can be helpful as its use seems to cause a sudden release of von Willebrand's factor into the bloodstream. After a 30 minute onset period, the use of DDAVP shortens the bleeding time for approximately 2 hours after the after DDAVP injection.

How can we manage Von Willebrand's disease?

There are two considerations;

1. DNA testing breeding animals so that this genetic disorder is not passed on. If you plan to breed your dog, von Willebrand's DNA testing is a good idea regardless of the breed but is a special concern for the "at risk" breeds.

2. And identifying and treating affected animals. If you own a member of one of the "at risk" breeds, consider having a screening test, especially if you are considering a major surgery.